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Tessier 30 with sublingual dermoid

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ABSTRACT

Tessier 30, also known as median midline cleft of the lower jaw, is a rare congenital anomaly presenting as cleft in the lower jaw that includes the tongue, the mandible, lower lip and the chin. There are less than 80 reported cases in the English medical literature since its discovery in 1819. We present one such case of Tessier 30 with presentation of cleft in chin and mandible along with ankyloglossia and a sublingual dermoid cyst that we reconstructed. 2 years post-operative follow up has been shown to be satisfactory.

Keywords: Dermoid; facial cleft; Mandibular cleft; Tessier 30; Tessier cleft.

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Case Report

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INTRODUCTION

Tessier 30 cleft or "median midline cleft"[1] was first termed by Dr Paul Tessier in 1976 but the features were first reported by Couronne in 1819. (Figure 1) Tessier 30 in the minor forms just affect the soft tissue of the lower lip but may extend to involve mandible, tongue, uvula, strap muscles of the neck, thyroid cartilage, clavicle and sternum. [2.3] Dermoids in the midline of neck have also been reported.[2] We present a case of Tessier 30 involving the chin, mandible, tongue along with a sublingual dermoid cyst between the mandibular processes. In this case we also show an interesting use of the cyst epithelium for intraoral reconstruction.

Case History:

A 3 year old boy presented with cleft of lower lip leading to incomplete lip seal during swallowing and tongue attached to floor of mouth. The clinical examination depicted midline cleft of the lower lip. Fusion at the mandibular symphysis was absent, leading to two mobile pieces with a diastema between the 7A, 8A and ankyloglossia. (Figure 2, Figure 3) Other systemic anomalies were ruled out. Surgical correction was planned under general anesthesia for bony and soft tissue repair in a single stage.

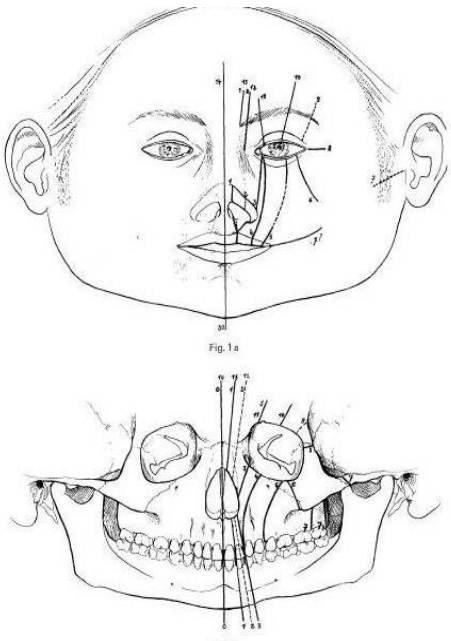

Figure 1: Classification of Tessier soft tissue and hard tissue clefts

Figure 2: Pre operative picture depicting clinical presentation of Tessier 30

Figure 3: Pre operative PA mandible showing the two separate mandibular processes

Figure 4: Marking of the tissue to be excised around the cleft and to release ankyloglossia

Figure 5: Sublingual dermoid exposed

Figure 6: Cyst epithelium opened and being pulled caudally.

Figure 7: Medialization of tissue to close the ventral surface of tongue. The white arrow shows raw area of the floor of the mouth and beyond being reconstructed by dermoid cyst epithelium.

Figure 8: Tongue and floor of the mouth reconstruction complete

Figure 9: Forceps holding the orbicularis oris muscle on the right side that needs to be sutured with its counterpart of left side to form complete oral seal. The cleft on chin after removal of soft tissue can also be appreciated here.

Figure 10: The finished reconstruction

Figure 11: 2 year post operative lip closure and scar

Figure 12: Complete freedom of tongue movements

Figure 13: PA mandible

The scar tissue around the cleft was marked and excised (Figure 4). Upon dissection, a sublingual dermoid cyst was found extending between the two mandibular segments. (Figure 5)

Release of ankyloglossia was performed. (Figure 5). Usually, a diamond shaped raw area; half on the ventral surface of tongue and half on the floor of the mouth were created. Usually, we medialize the edges for closure which post operatively result in scar contracture and loss of tongue freedom to some extent.

Here, traditional linear closure of the ventral aspect of tongue was performed. It was evident that after excision of scarred cleft tissue and release of ankyloglossia, the remaining soft tissue was inadequate.(Figure 6) The cyst fluid was drained and epithelium was spread inside out and used to augment the floor of the mouth, alveolar mucosa on the labial side, labial sulcus. (Figure 7) It mucosalized fast. (Figure 8)

The mandibular cleft was explored with minimal periosteal stripping. Segments were aligned at lower border and fixed in reasonable occlusion with trans-

osseous wiring. (Figure 13) Labially, the continuity of orbicularis oris was restored first and intraorally

lengthening of tissue was not required for closure since cyst epithelium was used (Figure 9). A Z-plasty was performed extra orally to match the white roll and vermillion of the lip and closure was performed with 6-0 prolene. (Figure 10)

Patient had an uneventful postoperative healing phase and achieved lip seal competence and showed free movement of tongue. The patient was on subsequent review cycle for 2 years. (Figure 11, Figure 12, Figure 13)

DISCUSSION

Reported by Couronne in 1819, "median midline cleft" of lower lip was first termed as Tessier 30 in 1976.[1] The clinical presentations may be just soft tissue involvement of lower lip or extend further as cleft in mandible[2] and/or ankyloglossia with/without a bifid uvula along with midline dermoids. [3] Often thyroid cartilage show underdevelopment. Widely spaced clavicles and bifid manubrium sterni may be associated with Tessier 30. Presence of presternal tags and ventriculo-septal defect in these patients are also documented.[2] Oostrom et al 1996, classified the Tessier 30 cases into soft tissue involvements, hard tissue involvements and associated anomalies depending on structures involved.[4]

First branchial arch develops in the 7th week of intrauterine life which further divides into the right and left mandibular processes that fuse in the midline to form the mandible. Severe hypoplasia of the mandibular process may give rise to the midline cleft or mesenchymal cells may invaginate between the mandibular processes and prevent forming of complete mandible.[4] Till 2015 only 80 such cases have been reported in the English medical literature- stating it as a rare congenital anomaly.[5]

The timing for surgery should be as early as possible[5] to prevent hypermobilty of segments, respiratory and feeding difficulty[6] but Armstrong, 1996, deferred the mandibular repair to 10 years of age to prevent damage to the developing permanent tooth buds.[3] Minor soft tissue cleft were corrected with "V" excision of cleft followed by a Z-plasty yielding a comparatively esthetic result as scar contracture is less.[3]

The surgical intervention is either single staged or multiple. In infants, the soft tissue closure performed first to help the patient form a complete lip seal for swallowing.[3]The ankyloglossia may be corrected at a later stage while performing the hard tissue repair.[3] Titanium plates fixation are preferred but require a removal procedure as in the symphysis region they may consequently retard the growth of the mandible.[5] Rao A.N.V, 2015, therefore emphasized on use of bioresorbable plates. Hard tissue repair often requires iliac bone graft or rib graft. When primary repair is performed in adults, further orthodontic and orthognathic correction becomes essential.[5] A series of soft tissue refining surgeries are required in cases of Tessier 30 cleft extending to the neck.

2 year post operatively our case shows no significant scar contracture or indicates need of revision. We used transosseuous wiring for the mandibular processes as it was cost effective, preserved the developing tooth buds and spared a secondary implant removal procedure. No major retrusion of mandible noted in postoperative period as we fixed the mandible at early age before the advent of major growth spurts. One of the objectives to achieve in these surgeries is to achieve adequate movement of the tongue. The freedom of tongue achieved at end of 2 years was also significant. (Figure 9)

CONCLUSION

Tessier 30 is a rare anomaly requiring an early diagnosis and intervention. Surgery, single stage or multiple, is the only answer to restore form, function and quality of life of these patients. Since the number of these cases are very less and presentations myriad, a standardized surgical protocol is lacking. Removal of scar tissue around the cleft and release of ankyloglossia often results in a tissue deficit. Reconstruction of the soft tissue in cleft region therefore requires careful planning, on-table innovation and novelty. Here we implement the use of dermoid cyst epithelium to reconstruct the floor of mouth and to provide adequate tongue movement.

Key Messages: Tessier 30 is a rare congenital anomaly. The restoration of form, function and quality of life is a challenge. These patients have severe ankyloglossia and ensuring the tongue freedom is a challenge. We use the dermoid cyst epithelium to achieve intra oral reconstruction. 2 years post op results yield a significant tongue movement and no significant extra oral scar

Conflicting Interest: Nil

Declaration of patient consent: The authors certify that we have obtained all appropriate patient consent form. In the form the patient has given her consent to use of her images and other clinical information to be published in scientific journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

REFERENCES

1. Tessier P. Anatomical classification of facial. Cranio-Facial and Latero-Facial Clefts. Journal of Maxillofacial Surgery.1976; 4:69–92.
2. Kawamoto HK. Rare craniofacial clefts. In: Mc Carthy JG, Plastic Surgery. W.B. Saunders; 1990. p. 2940.

3. Armstrong AP, Waterhouse N. Tessier 30 Median mandibular cleft: A case report and literature review. Br J of plast surg 1996; 49:536-8.
4. Oostrom CA, Vermeij-Keers, Gilbert PM, Van Der Meulen JC. Median cleft of lower lip and mandible: Case reports, a new embryologic hypothesis and subdivision. Plast Reconstr J 1996; 97:313-20.
5. Rao A.N.V, Complete Midline Cleft of Lower Lip, Mandible, Tongue, Floor of Mouth with Neck Contracture: A Case Report and Review of Literature, Craniomaxillofacial Trauma and Reconstruction Vol. 8 No. 4/2015
6. Bhattacharyya NC, Kalita K, Gogoi M, Deuri PK. Tessier 30 facial cleft. J Indian Assoc Pediatr Surg 2012;17:75-7.
7. Ali AAA, Tessier Number 30 Median Mandibular Cleft With Congenital Heart Anomalies in Qena, Egypt, The Cleft Palate-Craniofacial Journal 56(2): May 2018